SECOND EDITION

RECONSTRUCTIVE PLASTIC SURGERY

VOLUME FOUR

CLEFT LIP AND PALATE CRANIOFACIAL DEFORMITIES

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RECONSTRUCTIVE PLASTIC SURGERY

Principles and Procedures in Correction, Reconstruction and Transplantation

VOLUME FOUR

CLEFT LIP AND PALATE CRANIOFACIAL DEFORMITIES

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RECONSTRUCTIVE PLASTIC SURGERY

CHAPTER 38

CLEFT LIP AND PALATE: INTRODUCTION

JOHN MARQUIS CONVERSE, M.D.,
V. MICHAEL HOGAN, M.D.,
AND JOSEPH G. MCCARTHY, M.D.

HISTORY

Each of the following chapters dealing with basic and clinical information to assist the plastic surgeon in the rehabilitation of the cleft lip-palate patient contains relevant historical remarks which, if repeated in detail, would be redundant. There is an excellent historical review of the subject of cleft lip and palate by Dorrance (1933) and another historical review by Rogers (1971). This introduction will be concerned only with establishing historical trends in the treatment of this congenital anomaly.

The Age of Empiricism

In their approach to the problem of cleft lip and palate, surgeons through the ages have attempted to correct the abnormal anatomical arrangement of the cleft lip and palatal tissues and achieve a "normal" appearance. In ancient times many congenital deformities, including cleft lip and palate, were considered to be evidence of the presence of an evil spirit in the afflicted child. These children were then often removed from the tribe or cultural unit and left to die in the surrounding wilderness.

Boo-Chai (1966) reported a case of successful closure of a cleft lip at approximately 390 A.D. in China, although the surgeon's name is not mentioned. In Europe many surgical techniques were used for the treatment of wounds during the early Christian era. Hot cautery was a special feature of Arabian surgery, whereas the scalpel was favored by Greek and Roman surgeons. Yperman (1295–1351) was a Flemish surgeon who appears to have written the first fully documented description of cleft lip and its surgical repair. He closed the freshened borders of the cleft lip with a triangular needle armed with a twisted wax suture, a common method of suture at the time. In order to approximate the internal and external wound edges, he reinforced the closure with a long needle passed through the lip some distance from the edges of the cleft; the needle was held in place by a wraparound figure-of-eight thread. A similar technique of lip closure was still being performed by Pancoast in 1844.

Palatal deformities caused by syphilis and gunshot wounds interested Jacques Houllier (cited by Gurlt, 1898), who appears to have been the first to propose direct suture of palatal perforations. However, the failure rate was high, and he suggested that, when surgery failed, the region could be occluded with wax or a sponge. Franco (1556) wrote: "... cleft lips are some-

times cleft without a cleft of the jaw or palate, sometimes the cleft is only slight, and at times the cleft is as long and as wide as the lip" (Rogers, 1967). In 1561 he wrote: "Those who have cleft palates are more difficult to cure; and they always speak through the nose. If the palate is only slightly cleft, and if it can be plugged with cotton, the patient will speak more clearly, or perhaps even as well as if there were no cleft; or better, a palate of silver or lead can be applied by some means and retained there" (Rogers, 1967). Palatal occlusion by plates of gold or silver was also described in 1564 by Paré, who designated such a plate as an "obturateur": Paré (1975) was also the first to use the term "bec-de-lièvre" ("harelip").

Tagliacozzi (1597) described a lip closure utilizing mattress sutures passed through all layers of the lip tissue. This was a departure from the prevailing technique of needle closure and figure-of-eight suture material reinforcement. Thus, in the sixteenth century, closure of cleft lip to improve appearance was widely practiced, and the need for closure of the cleft palate to improve speech was appreciated in more limited surgical circles.

Treatment of the protruding premaxilla using a head bandage to achieve external compression of the premaxillary segment, thereby reducing it to a more favorable position for lip closure, was introduced by Desault and Bichat (1798). Over the years, various combinations of intraoral and extraoral devices were developed in order to reduce the protruding premaxillary segment and also to maintain the lateral arch segments in adequate anatomical relationship with the lower jaw. At the present time, these efforts appear to have reached their latest stage of development in the sophisticated work of Georgiade and Latham (1973).

The origins of the present techniques for successful closure of the secondary cleft palate are found in the early work of von Graefe and Roux, who in 1816 and 1819, respectively, closed the cleft of the *soft palate* with interrupted twine sutures. In Roux's patient, a dramatic change in the patient's voice was immediately noted and described.

Direct closure of the *hard palate* followed in 1826. Dieffenbach recommended that clefts of the hard palate could be closed by separating palatal mucosa from the bone. While he also recommended lateral relaxing osteotomies to close clefts of the secondary palate, he did not employ these until 1828. This technique is still employed in certain centers at the present time.

Early closure of the soft palate to induce a narrowing of a wide cleft of the hard palate was mentioned in 1828 by John C. Warren of Boston. This approach to wide clefts of the hard palate was repopularized by Schweckendiek in 1962 and is currently the subject of much debate (Fára and Brousilova, 1969). Langenbeck in 1859 and 1861 emphasized the need to elevate the periosteum with the palatal mucosa, thus forming bilateral mucoperiosteal flaps. This flap technique is still in use in some centers today. Veau drew attention to the fact that palatal lengthening was not achieved by this technique, launching a full-scale attack on the technique in the Deutsche Zeitschrift für Chirurgie in 1936 (Converse, 1962). He converted Langenbeck's bipedicle flaps into single pedicle flaps based on the descending palatine vessels. Modifications of Veau's basic techniques were made by Wardill (1937), Kilner (1937), and Peet (1961), resulting in a push-back technique for closure of clefts of the secondary palate that is widely used today. Simultaneous lengthening of the nasal surface of the velum can be accomplished by the Cronin modification (1957) (see Chapter 45).

Mirault introduced the modern cross-flap technique of lip closure in 1844, and since that time nearly every conceivable type of flap—triangular, rectangular, or curvilinear—has been tried. Mirault's technique remained popular and was advocated during the twentieth century by Blair and Brown (1930). Further modification of cleft lip closure was described in 1884 by Hagedorn, who devised a rectangular flap technique to prevent linear contracture. This procedure appears to have led to the operation of LeMesurier in 1949. During this period Z-plasty techniques were also used in various guises to relieve the tendency to linear scar contracture. This line of endeavor led to the Tennison (1952) low triangular flap technique and the high Z-plasty–rotation flap of Millard (1958) (see Chapter 43).

Throughout the evolution of the techniques of treatment for cleft lip and palate, therapy for ancillary problems such as dentoalveolar arch deformities, nasal abnormalities, maxillary hypoplasia, and speech difficulties had also progressed to a point where, in modern times, teams of specialists have been formed to manage the total problem, grown too complicated for one or two disciplines alone.

Management of the arch deformity in cleft palate by techniques of banding and prosthetic stabilization failed to achieve the goal of an adequate upper-lower dental arch relationship in

early approaches to this problem. Orthodontic therapy proceeded during the period of the eruption of the permanent teeth, usually during the period of mixed dentition, and often after years of treatment, a Class III malocclusion with significant crossbite remained (see Chapter 49).

This led dental innovators such as McNeil (1954) and Burston (1958) to advocate orthodontics in the first year of life in an attempt to establish proper arch relationships. They postulated that early alignment of arch segments would aid normal development of the maxilla. Arch position was maintained by appliances, initially a combination of internal and external appliances and finally a simple internal appliance. However, removal of the retaining appliance before puberty often resulted in recurrence of the original arch deformity. It was then thought that perhaps primary bone grafts might (1) stabilize the arch and (2) either grow or promote growth of the uninhibited maxilla. These speculations, however, had no scientific basis (see Chapter 48).

The effectiveness of primary bone grafting in the treatment of cleft palate arch deformities has not been satisfactorily determined. Initially, surgeons attempted bone graft in the region of the incisive foramen in an effort to improve their statistics on successful palatal closure (Lexer, 1908; Drachter, 1914). To produce adequate bony continuity between the premaxilla and lateral bony segments appeared to some surgeons such as Axhausen (1952) as the "final problem in the repair of complete clefts at the present time." The mere presence of the bony gap was enough to inspire a general surgical rush to fill it. However, as will become apparent in Chapter 48, filling the gap was not the end of the matter. Bone grafts appear to be unable on their own to "hold apart" any arch that has a tendency to collapse; the bone graft absorbs under pressure. Nor do primary bone grafts grow as was originally postulated, but rather they hinder growth with a significant limitation of maxillary development and a dramatic increase in crossbite malocclusion and pseudoprognathism (Kling, 1964).

Moreover, as the story of primary bone grafting in cleft palate surgery unfolds, it tends to confirm the prescience of Pruzansky, who in 1964 condemned the unscientific and unsubstantiated use of primary bone grafting when bone graft fever was sweeping many surgical circles.

In retrospect, however, we must marvel at the ingenuity of surgeons of the past who made major progress utilizing the trial and error method in an era when corollary scientific information was virtually nonexistent. There were, however, surgeons throughout history who attempted to apply their knowledge of anatomy and physiology and use scientific discipline in the design of their surgical procedures. The anatomical observations of Pancoast (1844) led him to design a specific operation, in which he divided the insertion of the palatal muscles "so as to prevent their straining the sutured edges of the palate assunder." Fergusson (1844–1845), noting that most palatal repairs disrupted, conducted a series of anatomical studies leading him to design an operation which divided the levator veli palatini muscles, the posterior tonsillar pillars, and, on occasion, the anterior tonsillar pillars. The incisions provided relaxation to the muscles and tissues of the palate in order to prevent lateral pull. The father of modern surgery of cleft lip and palate, Victor Veau, spent many long hours studying embryologic specimens. His contributions to the study of cleft lip and palate in and outside of the operating room are significant.

The Scientific Approach

In surgery of the cleft lip, Veau (1931, 1938) pointed out the paucity of muscle fibers in the medial aspect of the unilateral cleft and also in the prolabial segment of the bilateral cleft lip:

> The median border of the cleft lip is sterile. This anatomic fact, the inadequacy of the musculature of the median aspect, should provide us with a surgical directive: Demand nothing from the inner aspect which is sterile, utilize to the maximum the muscles of the lateral aspect which is fertile, sacrifice all of the mucosa of the inner aspect, but preserve carefully all of the mucosa of the lateral aspect.
>
> The principal cause of the mediocre results obtained in bilateral cleft lip repair is the absence of muscle in the prolabial segment of the lip. One can hope for contour and shape approaching the normal only if the lip contains muscle. I have long emphasized this fact: The muscular sterility of the prolabial segment.

In the treatment of the bilateral cleft lip, Veau was one of the first to allow the pressure of the repaired lip to recess the premaxilla.

> We are operating on faces in full evolution. The profile of the face will be submitted to a dual transformation. In the nose, the vomer will grow on condition that it has not been altered and it will increase the projection of the nose. In the lip, the reconstituted muscular ring in front of the premaxilla will push it backward. The operation of the cleft lip in the newborn is not an ordinary definitive operation of the type one does in plastic surgery in the patient in

whom growth is completed. Our role, in the newborn, is to create conditions of development as close to the normal as possible.

In condemning surgical intervention upon the vomer to recess the premaxilla, Veau wrote:

In order that the face of the newborn becomes a normal adult face, a series of unknown factors must come into play. All of these factors have their role in the distribution of forces which create the definitive form. They are the instrumental contributions the assembly of which makes the harmony we know. Eliminate the violins and you will no longer recognize a Beethoven symphony. That is what we have done (by sectioning the vomer) in the treatment of bilateral cleft lip: We have done away with the axial beam supporting the evolution of the face.

It was in embryology, however, that Victor Veau made his greatest contribution. His career as an embryologist started when he was over 60 years of age.

I am only a surgeon, yet circumstances have led me to play the role of an embryologist.... Yesterday, everyone said "cleft lip is caused by the absence of coalescence of the processes of the face." Tomorrow, they will say, "cleft lip is caused by the persistence of the subnarial epithelial membrane."

This concept is not my own; it is the concept of Professor A. Fleischmann, who is still living in Erlangen, where he spent his entire academic career as Professor of Zoology. I have been, however, the gardener who has been responsible for the growth of the small plant, once it was germinated. The embryologists ignored Fleischmann, or only referred to his hypothesis with irony. I showed that Fleischmann's hypothesis could be applied to all clinical varieties of the cleft lip malformation and, in addition, I have supported the hypothesis by embryological findings outlined in drawings of the stages of development of the subnarial region.

I would like to relate the set of circumstances that led me to explore an area that was not my own. Until 1930, I had never looked at an embryo. I knew of the development of the embryo only by what is found in books. I was searching for an operative method for the treatment of cleft lip and I was trying out various methods; I ascertained the fact that the only productive methods were those which approximated normal development; surgery of malformations is experimental biology. In 1926, I wrote a paper on "The role of the prolabial segment in the formation of the face." The theory of the coalescence of the processes led me to a method that I thought to be a good one because it had an embryological basis. I experienced a series of disasters. I was deeply distressed. What was wrong? Was it the surgical technique that was not successful or was embryology providing the wrong directives? I did not understand that I should look at the embryo as a surgeon instead of searching for new ideas in surgical techniques. I was encouraged to go to Vienna to see Professor Fischel who had the famous collection of embryos. There I heard the name of Professor Fleischmann and I began to have some precise idea of the evolution of the face.

The ideas of Fleischmann tallied with what I know of the various types of the cleft lip malformation; but I had difficulty in understanding the work of the German author; his pictures were poorly demonstrative. I wrote to him asking for explanations. Since that time, we have not ceased to be in touch with each other. We have written volumes of letters to each other.

In 1935, I wrote a paper entitled: "Hypothesis of the initial malformation of the cleft lip." I did not try to do the work of an embryologist. Staying on clinical grounds, I showed that the theory of the facial processes fitted poorly with what I observed in the cleft lip; the theory of Fleischmann, on the other hand, appeared to be the key to all the anatomical details and clinical varieties of the deformity. This was an indirect attack on the classical theory. Fleischmann had sent me diagrams drawn from cat embryos for this paper. These drawings were necessary, I felt, to provide a visual explanation of the theory of the professor from Erlangen.

I sent this paper to Professor Hochstetter, whom I did not know. I admired his work; he had been the first to describe the oronasal membrane, which is an incomprehensible finding according to the theory of the facial processes. Hochstetter did not go as far as to denounce the theory of the facial processes, but his own research, in addition to what I had observed in Fischel's laboratory, convinced me that the facial process theory was a "myth" that has vitiated the study of embryology of the face.

Hochstetter answered my letter, "I have had two specimens of cleft lip embryos put away in a drawer for many years; I have never discussed these specimens because I do not understand them. I am sending them to you." You can imagine how joyful, but at the same time, how anxious, I was when I looked at these specimens. There I found the indisputable proof of the Fleischmann theory. These specimens were embryos of 22 mm (unilateral cleft) and 23.3 mm (bilateral cleft).

I then returned to Vienna. With Hochstetter, I discussed the embryos at great length and in great detail. In Fischel's laboratory, I worked with his first assistant, Professor Politzer. We wrote a paper entitled: "The primary palate. Formation. Anomalies." This is a work of pure embryology; we studied 140 embryos from 5 to 25 mm in size at which the definitive form of the face is constituted.

While I was working on this paper (April, 1935), I went to Heidelberg to operate on cleft palate patients in the service of Professor Kirschner. I visited the embryological laboratory of Professor Keibel, who had just recently died. I arrived when they were finishing the staining of a specimen of a 22 mm embryo with a cleft. It showed renewed proof of Fleischmann's theory. Professor Hoepke, the first assistant, to whom I explained my idea, was not convinced.

Embryologically, the oronasal membrane which plays a role in the cleft lip is constituted by two fundamentally different formations: (A) The floor of the nose between the integument and the nasopalatine canal. This region is formed by the primary palate, a very precocious embryonic structure (5 mm, 2nd week) which appears when the mesoderm has invaded the epithelial wall (7 mm) and is definitively constituted when bone has commenced to differentiate into the undifferentiated mesenchyme (11 mm, 5th week). (B) The hard and soft palate. This long partition is constituted by the secondary palate, a relatively late embryonic formation, definitively constituted when the palatine processes have achieved their fusion (30 mm, 12th week). The malformation in the secondary palate is the congenital cleft of the palate. Most often (6 out of 10) the malformation of the primary palate, the true cleft lip, is associated with a malformation of the secondary palate and the two deformities form a teratologic entity which is dis-

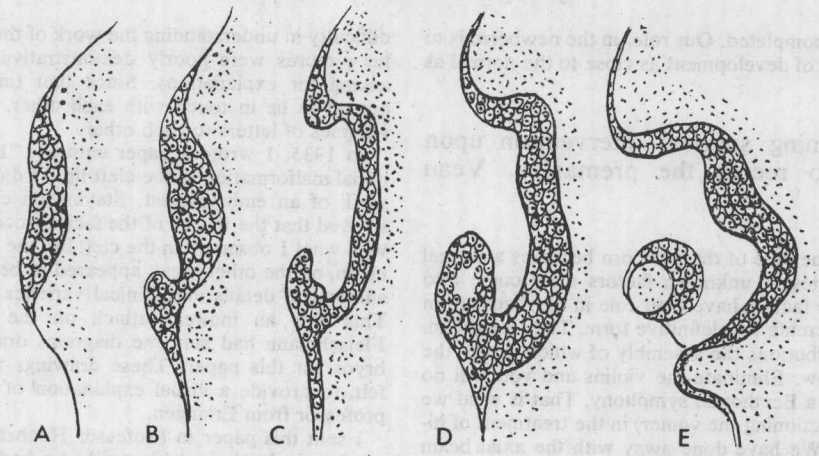

FIGURE 38–1. Formation of the primary palate. *A*, The plaque is the initial stage. It is formed by the localized thickening of the ectodermal covering (5-mm embryo). *B*, The fossa is produced by the raising of the edges of the plaque and the formation of a spur on the caudal aspect. *C*, The epithelial wall is the result of the increase in the height of the spur by the drawing together of the edges of the fossa. *D*, The disappearance of the wall coincides with the spread of the mesoderm. It is not possible to say whether the primary role is played by the ectoderm, which becomes hollow, or by the mesoderm, which perforates it. *E*, The primary palate is formed by the extension of the mesoderm, whose progressive growth leads to the formation of the subnarial region (16-mm embryo). (Redrawn after Veau, V., and Politzer, J.: Le palais primaire. Formation. Anomalies. Ann. Anat. Pathol., *13*:275, 1936.)

sociable because of its embryonic origins, but which forms, nevertheless, a clinical and surgical entity.

The gist of Fleischmann's hypothesis consists in the following: The cleft palate is the arrest of the disappearance of the epithelial membrane which remains intact, not penetrated by the adjacent mesoderm. Figure [38–1] is the diagram which Politzer and I arrived at in 1936. It summarizes the formation of the subnarial region. The legends explain the 5 stages. We used 108 drawings of normal embryos to represent these stages. We made an effort to eliminate the role of our imagination. We avoided making any comments on our illustrations for fear that these comments might be prejudiced. We did not allow ourselves to define the process of evolution. . . . In our paper the word "process" does not appear.

We have quoted the work of Victor Veau at length in order to demonstrate his awareness of other scientific disciplines and his dedication to scientific objectivity. He employed information outside of his own narrow field. His style and approach can be an inspiration to those who continue to manifest an interest in cleft lip and palate.

Fára's study of the anatomy of cleft lip and palate (1968), Kriens' (1969) research on the anatomy of the cleft palate and velopharyngeal region, the investigation by Lubker of the physiology of the velopharyngeal mechanism (1968), Warren and Devereux's work on the aerodynamics of the velopharyngeal region (1966), the acoustical analysis of speech and velopharyngeal incompetence by Isshiki and coworkers (1968), the embryologic studies of Avery (1961), the anatomical studies of Stenström and Öberg (1960) on cleft lip–nose deformity, and the analysis of the anatomy of the columella by Latham (1970) provide the kind of information which must be sought if we are to continue the strong heritage of the past in seeking our final goal, to make the abnormal as normal as possible.

Contemporary Theories

Throughout the historical development of the treatment of cleft lip and cleft palate, different aspects of the problem have alternately received priority. At the time of publication of this book, there appears to be an emphasis on the problem of crossbite and malocclusion following cleft lip and palate surgery (see Chapter 42).

Treatment of the anterior palatal deformity has been modified following the publication by Walker, Collito, Mancusi-Ungaro, and Meijer (1966) of data indicating the deleterious effect of extensive lateral undermining to facilitate the lip repair. The authors suggested that utilizing the technique of lip adhesion, followed in several months by lip closure without lateral periosteal or soft tissue undermining, significantly reduces the incidence of crossbite and malocclusion.

In addition, early complete closure of the primary and secondary palates can also produce significant dental deformities. Ross and

Johnston (1972) suggested that surgery should not be performed on the hard palate in areas adjacent to or abutting on teeth. As an alternative approach, Lindsay (1974) advocated a simple closure of the soft palate with obturation of the hard palate until age 2 or 3 years, when closure of the hard palate is completed by one of the "less" periosteally disturbing procedures, such as the von Langenbeck palatoplasty.

The effect of the later approach on speech development is not clear. In the past few decades our primary concern has been for speech, and we have been comfortable in the knowledge that if we gave something away in terms of initial dental management in order to obtain normal speech, it could be recouped later by advanced orthodontic techniques. However, it has now become evident that it is preferable *not* to yield on early dental management in order to decrease dental deformities; we must also be doubly alert to the effects of this "new" approach on speech. Speech compromises in early childhood might prove more difficult to correct later on than dental ones.

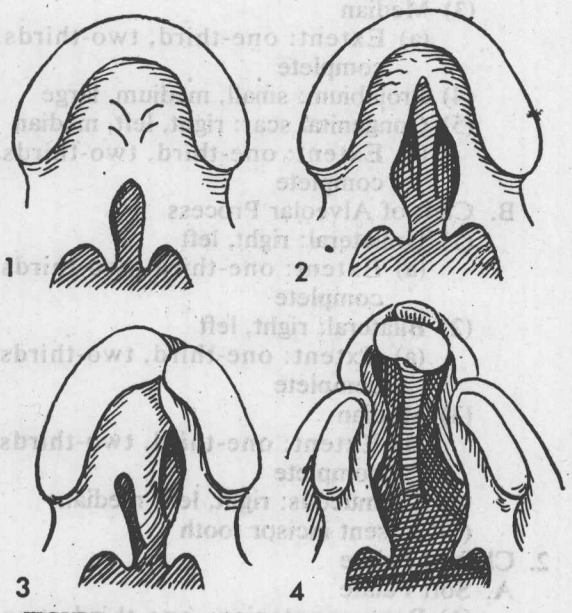

FIGURE 38–2. The Veau classification of the clefts of the lip and palate. Group 1: cleft of the soft palate only. Group 2: cleft of the soft and hard palate as far forward as the incisive foramen. Group 3: complete unilateral alveolar cleft, usually involving the lip. Group 4: complete bilateral alveolar cleft, usually associated with bilateral clefts of the lip. (After Veau, 1931.)

CLASSIFICATION

While various classification systems have been proposed, only a few have found wide clinical acceptance.

In the classification of Davis and Ritchie (1922), congenital clefts were divided into three groups according to the position of the cleft in relation to the alveolar process:

Group I, prealveolar clefts, unilateral, median, or bilateral

Group II, postalveolar clefts involving the soft palate only, the soft and hard palates, or a submucous cleft

Group III, alveolar clefts, unilateral, bilateral, or median

Veau (1931) suggested a classification divided into four types (Fig. 38–2):

1. Cleft of the soft palate only.
2. Cleft of the hard and soft palate extending no further than the incisive foramen, thus involving the secondary palate alone.
3. Complete unilateral cleft, extending from the uvula to the incisive foramen in the midline, then deviating to one side and usually extending through the alveolus at the position of the future lateral incisor tooth.
4. Complete bilateral cleft, resembling type 3 with two clefts extending forward from the incisive foramen through the alveolus. When both clefts involve the alveolus, the small anterior element of the palate, commonly referred to as the premaxilla, remains suspended from the nasal septum.

Kernahan and Stark (1958) recognized the need for a classfication based on embryology rather than morphology. The roof of the mouth—from the incisive foramen or its vestige, the incisive papilla, to the uvula—is termed the secondary palate. It is formed after the primary palate (premaxilla, anterior septum, and lip). The incisive foramen is the dividing line between the primary and secondary palates (Fig. 38–3).

A cleft of the secondary palate is further classified as incomplete or complete, depending upon its extent. An incomplete cleft is the common cleft of the velum, while a complete cleft includes both the velum and the hard palate as far as the incisive foramen. To this classification must be added the cleft of the mesoderm of the palate, or submucous cleft, which may be camouflaged unless the uvula is cleft (see Chapter 45). While it may not be easy to

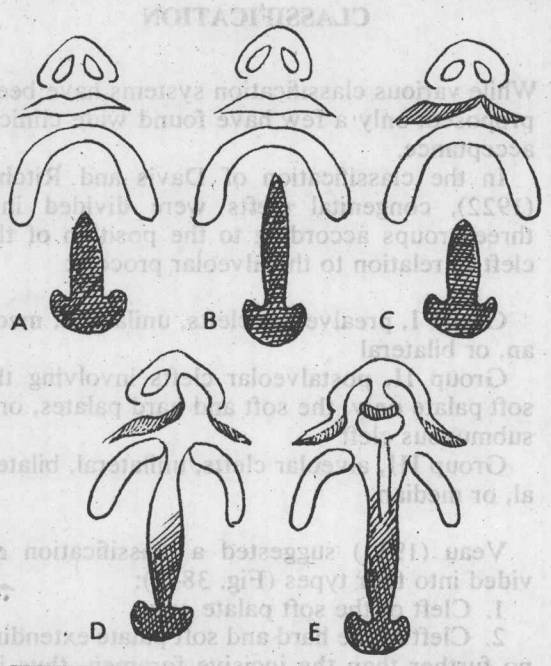

FIGURE 38–3. Classification of cleft palate. The division between primary palate (prolabium, premaxilla, and anterior septum) and secondary palate is the incisive foramen. *A*, Incomplete cleft of the secondary palate. *B*, Complete cleft of the secondary palate (extending as far as the incisive foramen). *C*, Incomplete cleft of the primary and secondary palates. *D*, Unilateral complete cleft of the primary and secondary palates. *E*, Bilateral complete cleft of the primary and secondary palates. (After Kernahan and Stark, 1958.)

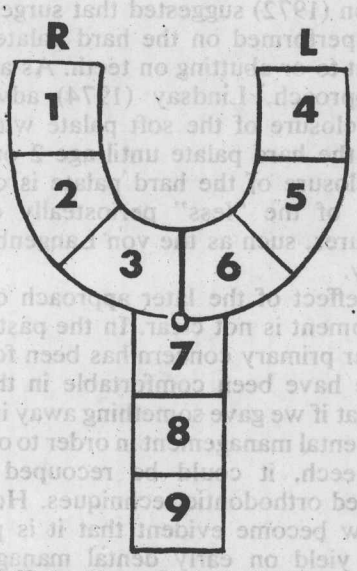

FIGURE 38–4. The striped Y classification. The involved area is filled in by pen and provides graphic demonstration of the site and extent of cleft involvement. (From Kernahan, D. A.: The striped Y—A symbolic classification for cleft lip and palate. Plast. Reconstr. Surg., *47*:469, 1971.)

detect dehiscence of the velum musculature, the presence of velopharyngeal incompetence and palpation of a notching of the posterior nasal spine aid in the diagnosis.

Kernahan (1971) subsequently proposed a striped Y classification (Fig. 38–4). As in the previous classification, the incisive foramen is the reference point. With stippling of the involved portion of the Y, the system provides rapid graphic presentation of the original pathology.

Harkins and associates (1962), at the instigation of the American Cleft Palate Association, presented a classification of facial clefts based on the same embryologic principles used by Kernahan and Stark. A modified version follows:

1. Cleft of Primary Palate
   - A. Cleft Lip
     - (1) Unilateral: right, left
       - (a) Extent: one-third, two-thirds, complete
     - (2) Bilateral: right, left
       - (a) Extent: one-third, two-thirds, complete
     - (3) Median
       - (a) Extent: one-third, two-thirds, complete
     - (4) Prolabium: small, medium, large
     - (5) Congenital scar: right, left, median
       - (a) Extent: one-third, two-thirds, complete
   - B. Cleft of Alveolar Process
     - (1) Unilateral: right, left
       - (a) Extent: one-third, two-thirds, complete
     - (2) Bilateral: right, left
       - (a) Extent: one-third, two-thirds, complete
     - (3) Median
       - (a) Extent: one-third, two-thirds, complete
     - (4) Submucous: right, left, median
     - (5) Absent incisor tooth
2. Cleft of Palate
   - A. Soft Palate
     - (1) Posteroanterior: one-third, two-thirds, complete
     - (2) Width—maximum (mm)
     - (3) Palatal shortness: none, slight, moderate, marked
     - (4) Submucous cleft
       - (a) Extent: one-third, two-thirds, complete

B. Hard Palate
   (1) Posteroanterior
      (a) Extent: one-third, two-thirds, complete
   (2) Width—maximum (mm)
   (3) Vomer attachment: right, left, absent
   (4) Submucous cleft
      (a) Extent: one-third, two-thirds, complete

3. Mandibular Process Clefts
   A. Lip
      (a) Extent: one-third, two-thirds, complete
   B. Mandible
      (a) Extent: one-third, two-thirds, complete
   C. Lip Pits: Congenital lip sinuses
4. Naso-ocular: Extending from the narial region toward the medial canthal region.
5. Oro-ocular: Extending from the angle of the mouth toward the palpebral fissure.
6. Oro-aural: Extending from the angle of the mouth toward the auricle.

Spina (1974) modified and simplified the above classification as follows:

Group I. Preincisive foramen clefts (clefts lying anterior to the incisive foramen). Clefts of the lip with or without an alveolar cleft.
   A. Unilateral
      (1) right { total when they reach the alveolar arcade / partial }
   B. Bilateral
      (1) total } on one or both sides
      (2) partial }
   C. Median
      (1) total
      (2) partial

Group II. Transincisive foramen clefts (clefts of the lip, alveolus, and palate).
   A. Unilateral { right / left }
   B. Bilateral

Group III. Postincisive foramen clefts.
   A. Total
   B. Partial

Group IV. Rare facial clefts.

Rare facial clefts are discussed and classified in Chapter 46.

EPIDEMIOLOGY

Clefts of the lip, with or without clefts of the palate (CL/P), must be distinguished from isolated clefts of the hard and soft palates (CP) because of different embryologic, etiologic, and epidemiologic backgrounds (Fogh-Andersen, 1942; Fraser and Calnan, 1961).

Clefts of the secondary palate can be induced in the mouse by teratogens after the primary palate has formed; moreover, clefting of the secondary palate in association with clefts of the primary palate probably represent a secondary (tongue positioning) rather than a primary defect (Trasler and Fraser, 1963).

Family studies have also shown that siblings of patients with CL/P have an increased incidence of CL/P but not of isolated CP; conversely, siblings of patients with CP have an increased frequency of CP but not of CL/P (Fogh-Andersen, 1942; Woolf, Woolf and Broadbent, 1963a,b).

The collection of epidemiologic data is associated with many problems. The surveys are conducted from three sources: birth certificates, hospital records, and habilitation or surgical records. The most accurate data are collected from the records of better hospitals. Birth certificates are often hastily completed and lacking in detail. Habilitative or surgical records tend to be slanted toward a certain segment of the cleft lip and palate population. Other factors which must be considered in evaluating the data include the percentage of ascertainment, the racial and socioeconomic make-up of the population segment under study, the quality of the records, and the absence of detail such as degree of clefting and presence of associated anomalies.

The reader is referred to the publications of Fraser (1970) and Ross and Johnston (1972) for a complete discussion of the epidemiology of cleft lip and palate.

Incidence. In the classic studies of Fogh-Andersen (1942), the overall frequency of cleft lip and palate in Denmark was reported as 1.47 per 1000 live births; the incidence of CL/P was 1.16 and that of CP was 0.34 per 1000 births. A similar overall incidence was reported by Woolf, Woolf, and Broadbent (1963a) for a section of the United States and by Wilson (1972) for a region in Great Britain. Racial differences will be discussed later in the chapter.

In most series cleft lip with palate involvement is reported as 1.5 to 3.0 times as fre-